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Successful Completion of Pregnancy in a Patient with a Hufnagle Valve

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PLACEMENT of the Hufnagle valve* is a useful therapeutic expedient in patients with aortic insufficiency, since more definitive surgical operation has not proved practical thus far. The following is a report of the first patient with such a valve in place to successfully complete a normal pregnancy.

REPORT OF A CASE

The patient, a 21-year-old white woman, had a normal infancy and childhood until the age of seven years, when, because of sore throats, nosebleeds and rheumatism, she was out of school for most of the next five years. Tonsillectomy was done during this initial illness.

When the patient was ten years old the family moved to California from South Dakota. Soon thereafter she had "swelling" of her body, and was out of school and sick most of the time for the next 18 months. At age 18 there was another similar episode, because of which the patient was in hospital two weeks, and was confined in bed at home for three months. From then until the time of this report she had no arthralgia, fever or major respiratory tract illnesses. Her feet occasionally swelled in warm weather, but not otherwise.

When first observed by the authors in November, 1955, she was able to do her own housework and shopping, but could not climb stairs without dyspnea. More than ordinary exertion at a slow pace was impossible. There was no exertional cough, hemoptysis or nocturnal dyspnea. The patient felt that her symptoms had increased, particularly in the preceding two years, and that she was more severely handicapped than previously.

The blood pressure was 155/40-0 mm. of mercury. There were no conjunctival or mucous membrane lesions, the color of the skin was normal, no abnormality was observed in the throat and there was no enlargement of lymph nodes. The teeth were sound. The lungs were clear to auscultation. Prominent pulsations were noted in the neck vessels, and a systolic thrill could be felt at the aortic area. There was a diastolic thrill at the mitral area. A loud to-and-fro aortic murmur was heard, which obscured the second sound. There was transmission of the sound downward along the left sternal border. A grade II systolic murmur at the mitral area, a diastolic rumble, and a snapping mitral first sound were heard. As determined by percussion, the heart was enlarged to the anterior axillary line. There was no enlargement of the liver. No abnormality was noted in the abdomen except for a prominent dynamic abdominal aorta and bounding femoral pulsations.

Fluoroscopically the lung fields were normal, as was the diaphragmatic movement. The left heart border was approximately 1 cm. from the left chest wall, and this appeared to be largely the result of left ventricular hypertrophy. There was very slight displacement of barium posteriorly in the oblique views. There was a dynamic aortic pulsation. The pulmonary segment was not prominent. The electrocardiogram showed a typical pattern of decided left ventricular hypertrophy. It was concluded that the patient had rheumatic heart disease, with aortic stenosis and insufficiency and associated mitral stenosis and insufficiency. Aortic insufficiency was considered to be the predominant lesion.

Both the patient and her husband wished to have definitive treatment. A Hufnagle valve was placed† in the descending aorta on April 23, 1956. Operation and the postoperative course reportedly were without event, and the patient made a normal convalescence. Shortly thereafter it became apparent that the patient was pregnant, and that conception may actually have preceded the surgical intervention.

The first and second trimesters of pregnancy were uneventful, the patient remaining quite comfortable, the valve sounds remaining unchanged, and vital capacity gradually increasing. There was a modest gain in weight. However, at the beginning of the eighth month, shortly after the Thanksgiving holiday, during which the patient had overeaten somewhat, there was an increase in blood pressure to 160/0 mm. of mercury and slight edema was noted at the ankles. Reserpoid and restriction of sodium intake were prescribed.

*The Hufnagle valve is a plastic sleeve containing a sliding ball valve, the whole unit being so designed that it can be clamped in place in the descending aorta below the arch. It prevents regurgitation of blood from the lower aorta, but of course does not eliminate that from the head and neck. The action of the ball valve makes a clicking sound.

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†By Dr. Henry Swan, after studies by Dr. Gilbert Blount in consultation.

Early on the morning of admittance to the hospital, December 17, 1956, the patient awakened acutely dyspneic and was aware of a decided change in the clicking sound of the valve. She was therefore put in hospital for further observation and treatment. The systolic blood pressure was then generally in the range of 180 mm. of mercury and the third phase remained at around 40 or 50 mm., with the fourth phase at zero. Digitalization was carried out, and with complete bed rest the patient seemed to respond satisfactorily. Most observers agreed that there was some change in the sound of the valve, which instead of having a clear-cut clicking sound, had a slight scraping sound before each click.

Upon vaginal examination the cervix was observed to be soft and effaced and slightly dilated. It was, therefore, decided to induce premature labor by rupturing the membranes. This was carried out, and an uneventful four-hour labor ensued, with normal delivery of a healthy child. The patient did extremely well immediately following delivery, but approximately 22 hours postpartum, she suddenly had a convulsive episode, became quite unmanageable and thrashed about on the bed, necessitating restraint. The blood pressure at the time was 300 mm. systolic. No diastolic reading was obtainable. The physician, thinking the valve might have stuck, rolled the patient on her side and pounded her back. More reserpine was given intramuscularly, and sodium amytal and magnesium sulfate also were administered to control the acute agitation. By the next day the patient was quite rational and oriented, but flaccid hemiplegia on the left was noted. No abnormalities were observed in the spinal fluid. An electroencephalogram was interpreted as showing spasm in the right temporal region. The hemiplegia cleared almost completely in the following week and the patient was discharged home.

It was the opinion of a neurosurgical consultant that cerebral-angiospasm was the most likely cause of the postpartum episode. Pre-eclamptic toxemia had been considered, but the absence of albuminuria or retinal changes was against this possibility. After conversation by phone with Dr. Hufnagle (designer of the valve) the physician attending the patient suggested the possibility of retrograde thrombosis from the valve with small cerebral embolization. No completely adequate explanation of the patient's postoperative complication has been evolved. At the time of this report, some six months after the episode after parturition, the patient was doing well. There was almost no residual hemiplegia, and the blood pressure was within the normal preoperative range at 140/0 mm. of mercury. The valve sound was normal.

SUMMARY

A woman in whom an artificial valve had been placed because of aortic insufficiency, subsequently had successful completion of pregnancy. Despite an unexplained postpartum complication the patient made satisfactory recovery.

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Osteochondrodystrophy (Morquio-Brailsford Type)

Occurrence in Three Siblings

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In 1929 Morquio[12] reported upon a family of five siblings, four of whom had the following abnormalities: The sternum "standing out to a point," relatively long limbs, genu valga, pes planus, deformity of the vertebral column, muscle weakness and atrophy, and peculiar gait resembling the walk of a duck. Radiographically there was a disturbance of osteogenesis of the epiphyses of all the bones. The children were of normal intelligence and normal pubertal development.

Three members of a family who had similar abnormalities are reported upon herein. These patients are of special interest first because they were of Japanese ancestry, which was not so in any previously reported cases, and secondly because two of the patients are twins, of which only one previous instance is known.[8]

The prognosis is generally considered poor but in cases where follow-up observations have been reported[8,13] this gloomy outlook has lacked support. Hirsch[8] from his observations of two families suggested the existence of a florid stage followed by a healing stage. One of the patients described in the present report (Case 1) seemed to have had a remission, but the period of observation thus far is not sufficiently long to refute the belief that the disease is progressive with death to be expected at an early age (14).

REPORTS OF CASES

Case 1. Patient A, a 14-year-old boy of Japanese parentage was first observed at home in December 1954. He had fallen out of his wheelchair and complained of pain in both knees. Roentgenographic examination of the legs showed greenstick fractures of both distal femora and both legs were put in long casts for a period of several months. At the end of this time firm bony union was found to have taken place. After the casts were removed, function of the legs slowly returned. In about six months he was able to discard cane and crutches and then was able to walk with a waddling gait.

From the medical history it was learned that the child was considered to be well by his parents until the age of seven when it was noted that he was of shorter stature than children of his age. At the age of ten years the patient was treated by a physician because of complaint of pain in the hips. By the age of 12 he was confined completely to a wheelchair

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